"STA-MCA bypass with encephalo-duro-myo-synangiosis combined with bifrontal encephalo-duro-periosteal-synangiosis" as a one-staged revascularization strategy for pediatric moyamoya vasculopathy

Esposito, Giuseppe ; Kronenburg, Annick ; Fierstra, Jorn ; Braun, Kees P J ; Klijn, Catharina J M ; van der Zwan, Albert ; Regli, Luca

Abstract: PURPOSE Moyamoya vasculopathy progressively compromises cerebral blood flow resulting in chronic hypoperfusion. The middle cerebral artery (MCA) territory and the bifrontal areas are the regions most frequently affected. Although most techniques aim to only revascularize the MCA territory, augmentation of blood flow of the bifrontal areas is of importance in the pediatric moyamoya population since these regions play an important role in cognition, intellectual development, and in lower extremity and sphincter function. We recently described a one-staged surgical procedure combining revascularization of three regions, the MCA territory unilaterally and the frontal areas bilaterally. The purpose of this article is to report our surgical experience in eight children and to emphasize the rational for bifrontal revascularization. METHODS We report a case series consisting of eight children where the following surgical strategy was applied: (1) a direct superficial temporal artery-to-middle cerebral artery (STA-MCA) bypass with encephalo-duro-myo-synangiosis (EDMS) for unilateral MCA revascularization; in combination with (2) a bifrontal encephalo-duro-periosteal-synangiosis (EDPS) for bifrontal revascularization. Patients' characteristics and 30-day follow-up data are reported. RESULTS The patient group consisted of six girls and two boys (mean age 10.0, range 4.2-17.5 years): six children presented with moyamoya disease, two with moyamoya syndrome. We performed a one-staged revascularization of one MCA territory and both frontal areas in all patients. No significant complications occurred. Two patients experienced postoperative focal seizures, successfully treated with anti-epileptic medication. CONCLUSIONS The single-staged STA-MCA bypass with EDMS combined with bifrontal EDPS allowed revascularization of three regions (the MCA territory unilaterally and the frontal areas bilaterally) and may serve as an alternative and safe treatment option for pediatric moyamoya patients.

DOI: https://doi.org/10.1007/s00381-015-2665-y

Childs Nerv Syst (2015) 31:765–772
DOI 10.1007/s00381-015-2665-y

TECHNICAL NOTE

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Received: 24 November 2014 / Accepted: 17 February 2015 / Published online: 27 February 2015

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Keywords Bypass · Cerebral revascularization · Children · Frontal area · Moyamoya · Pericranial flap

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Introduction

In patients with moyamoya vasculopathy (MMV), surgical revascularization is performed to prevent recurrent stroke and to stabilize or improve neurological outcome and quality of life [1]. Applied techniques of revascularization vary, and it remains unclear whether one technique should be preferred over the other, both in adults and in children [1–3]. The most commonly used direct revascularization procedure is the

superficial temporal artery to middle cerebral artery (STA-MCA) bypass that instantly augments blood supply to the middle cerebral artery (MCA) territory. Indirect techniques rely on the overlay of vascularized tissue (like the temporal muscle, pericranium, and dura) onto the cerebral cortex in order to promote neoangiogenesis over time [1–3]. Combined revascularization procedures provide the advantages of both techniques [2–7].

In addition to symptoms that can be ascribed to the MCA territory, children may present with lower extremity motor weakness and neuropsychological dysfunctions probably due to involvement of the frontal lobes [8–12]. In pediatric MMV, the cerebral blood flow (CBF) in the bifrontal areas as well as in the anterior watershed territory may continue to worsen despite good collateral formation or successful revascularization of the MCA territory [9–12]. Therefore, it is important to consider timely revascularization of the frontal areas, to prevent neurocognitive decline in pediatric patients [8–12].

We recently published the technique of a one-staged surgical procedure for children diagnosed with MMV, which combines revascularization of three regions, the MCA territory unilaterally and the frontal areas bilaterally. This technique consists of unilateral STA-MCA bypass with encephalo-duro-myo-synangiosis (EDMS) and bifrontal encephalo-duro-periosteal-synangiosis (EDPS) [3].

The purpose of this article is to report our surgical experience in a series of eight pediatric moyamoya patients and to emphasize the rational for bifrontal revascularization.

Patients and methods

Patients

We retrospectively reviewed all pediatric patients with MMV who were operated on by performing the one-staged procedure (unilateral STA-MCA bypass with EDMS and bifrontal EDPS) between December 2009 and March 2012.

Preoperative work-up includes magnetic resonance imaging (MRI), six-vessel digital subtraction angiography (DSA), H_2O-positron emission tomography (PET), and neuropsychological evaluation. These investigations are repeated 1 year after surgery.

According to our protocol, "unilateral STA-MCA bypass with EDMS combined with bifrontal EDPS in one-step surgical procedure" was indicated for children presenting with both the following conditions: (1) hemodynamic compromise (impaired CBF and/or cerebrovascular reserve—CVR) or ischemic symptoms in a MCA territory; (2) hemodynamic compromise (impaired CBF and/or CVR) or ischemic symptoms in both frontal regions.

We registered clinical characteristics (disease, sex, age, preoperative symptoms), surgical data (side and type of surgery, operative time), and postoperative course and events (occurrence of complications, days of admittance, 30-day follow-up).

Unexpected events within 30 days after surgery are described as perioperative complications. Episodic symptoms within 30 days postoperatively which resembled preoperative events in terms of frequency, duration, and aspect (e.g., unchanged transient ischemic attacks (TIAs)) were not included as perioperative complications.

We report in details the treatment and follow-up data of a child affected by moyamoya disease (MMD).

Description of the surgical technique

Direct (STA-MCA) and indirect (EDMS) bypass for unilateral MCA territory revascularization

The patient is placed in supine position with the head mildly extended in the 3-points Mayfield headrest and 30° rotated to the opposite side. Care has to be paid not to injure the contralateral superficial temporal artery (STA) as this would compromise a future STA-MCA bypass on the opposite side. We do not shave the hair.

The incision starts over the parietal branch of the STA. The STA-MCA bypass is performed according to the classic technique described elsewhere [13]. In brief, the parietal branch of the STA branch is dissected and prepared under microscopic view: this donor vessel is kept intact up to the anastomotic procedure. The temporal muscle is cut along the skin incision and a craniotomy is performed on the Sylvian point over a large cortical MCA branch. The dura mater is opened in a star-shaped fashion preserving the main branches of the middle meningeal artery. After meticulous hemostasis, the dural flaps are reflected subdurally under the bone window, to obtain encephalo-duro-synangiosis (EDS) (Fig. 2a–b). The cortex is inspected for the largest cortical M4 recipient artery, which is dissected by means of arachnoid opening. A segment with no or only few cortical side-branches is chosen (1–2 of the tiny side-branches may need to be interrupted). Two silicon triangle-shaped background sheets are inserted beneath the recipient artery, in order to facilitate the construction of the anastomosis. A temporary nontraumatic microvascular clip is placed across the exposed STA proximally. The distal STA is cut in a fish-mouth to increase the opening diameter of the donor vessel and prepared for the micro-anastomosis. A blue dye is applied onto the donor and recipient vessels to improve visualization during the anastomotic procedure. Nontraumatic temporary microvascular clips are applied on the recipient vessel. A linear arteriotomy on the

cortical recipient is performed so that the micro-anastomosis is at least 2.5 times the size of the diameter of the recipient vessel. Two 10-0 monofilament sutures are applied at the toe and the heel of the anastomotic site to anchor the donor and recipient vessel. The micro-anastomosis is performed with interrupted 10-0 monofilament sutures to allow anastomosis growth with time. Before knotting the last suture, the anastomosis is flushed to clear air. Flow is reestablished by removing first the distal and then the proximal temporary clips on the cortical MCA recipient artery, and finally the clip on the proximal STA (Fig. 2c). Bypass patency is assessed with Indocyanine green video angiography (performed by the use of a commercially available microscope, OPMI® Pentero, Carl Zeiss Co., Oberkochen, Germany). The flow in the bypass is quantitatively assessed using an intraoperative flow probe (Transonic Systems Inc., Ithaca NY). Flow values over 15 ml/min are considered sufficient. In pediatric moyamoya patients, flow values are expected to be between 15 and 50 ml/min, depending on patients' age, donor and recipient characteristics, and hemodynamic conditions. The micro-anastomosis is observed for 20 min to make sure there is no decrease in flow. Closure is performed by covering the exposed cortex with the temporal muscle and by suturing the muscle to the dural edges (to obtain encephalo-myo-synangiosis—EMS). The bone flap is then secured into place above the muscle. Attention is paid to avoid any compression of the bypass (Fig. 2f).

Bifrontal EDPS

After completing the direct (STA-MCA) and the indirect (EDMS) bypass for a unilateral MCA territory revascularization, the skin incision is extended frontally 4 cm over the midline, staying behind the hairline (Figs. 1a and 2a). This extension can also be made in a zigzag fashion [14]. The scalp flap is reflected anteriorly and a vascularized bifrontal pericranial flap, consisting of the periosteum and the overlying loose areolar layer, is dissected (Figs. 1a–b and 2d). The pericranial flap is kept pediculated towards the biorbital regions to maximize vascular supply. This pediculated pericranial flap will serve to perform the bifrontal encephalo-periosteal-synangiosis (EPS). Two separate frontal parasagittal craniotomies (4×5 cm) are

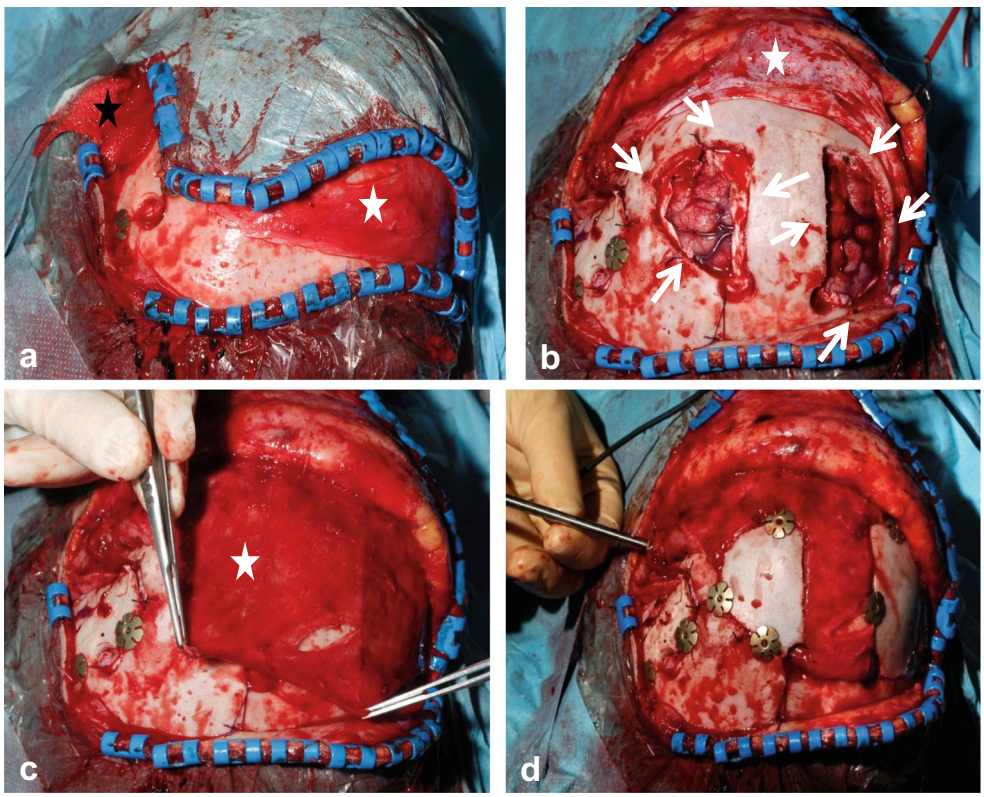
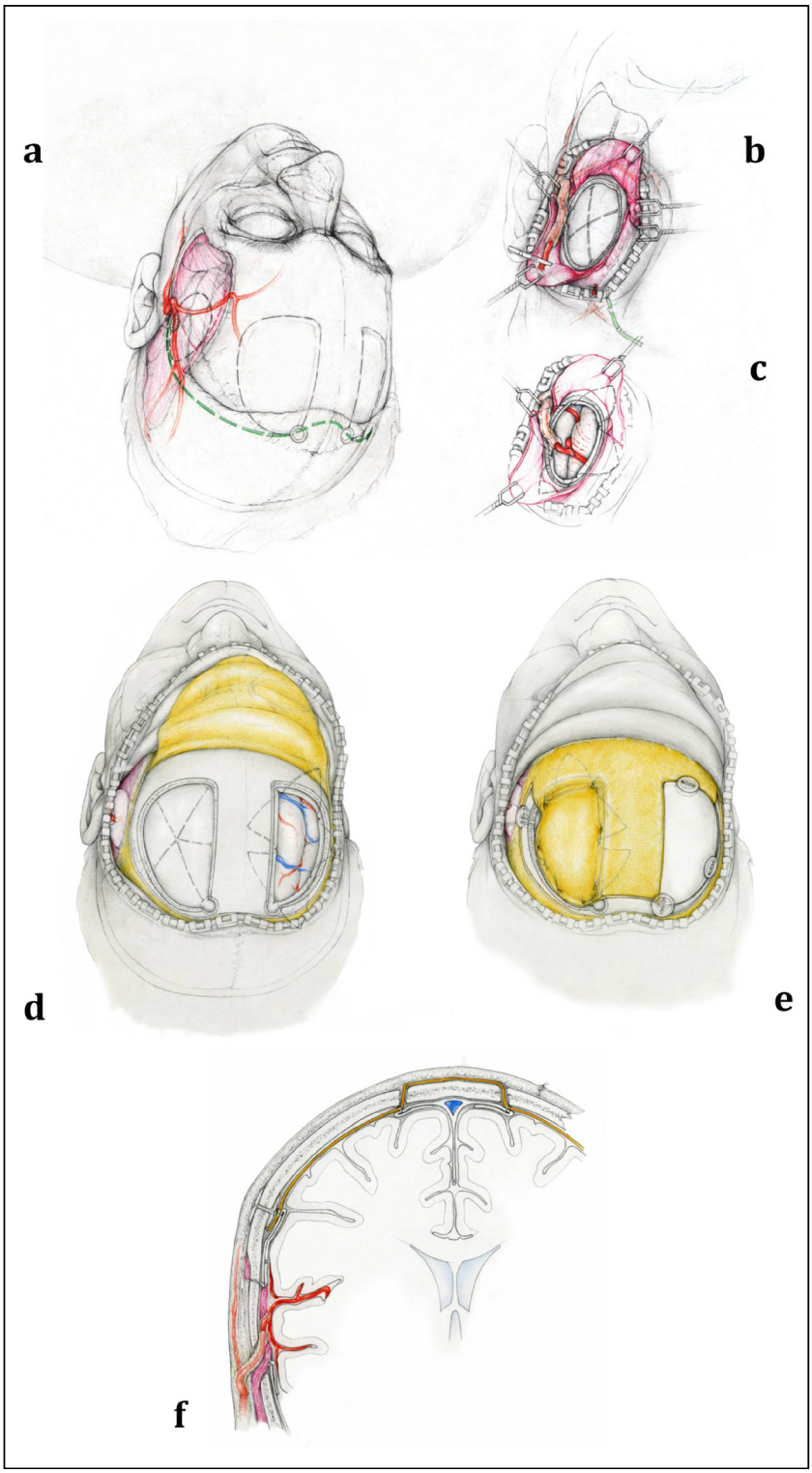


Fig. 1 Intraoperative pictures: **a** After having performed a STA-MCA bypass and EDMS on the left side (the *white arrow* indicates the craniotomy and the *black asterisk* a cottonoid covering the bypass), the skin incision is extended 4 cm over the midline behind the hairline. The dissected frontal pericranial flap is indicated by the *white asterisk* (*no-shaving technique*). **b** The scalp flap is reflected anteriorly; the vascularized frontal pericranial tissue is dissected and reflected on the scalp flap (see *white asterisk*); two symmetric bilateral frontal parasagittal craniotomies are performed; the underlying frontal dura is then opened in a star fashion, and the dural flaps are inverted onto the cortex around each frontal bone window, to obtain EDS (see *multiple white arrows*). **c** The pericranial flap (white asterisk) is placed over the cortex surface and is sutured to dural edges to obtain EPS. **d** The bone flaps are repositioned



performed: one on the left and one the right side. The craniotomies are localized 2 cm away from the midline to avoid injuring the superior sagittal sinus (SSS) and the parasagittal veins. The dura is then opened in a star-shaped fashion, and, after meticulous hemostasis, the dural flaps are inverted and reflected under the edges of each frontal bone window to obtain EDS (Figs. 1b, and 2d). Small cortical arachnoidal openings are made. The periosteal flap is then positioned over the cortical convexity and sutured laterally to the dura (EPS) (Figs. 1c and 2e). Both frontal bone flaps are repositioned and fixed (Figs. 1d and 2e–f). The scalp flap is re-approximated

◀ **Fig. 2** Sketch of the proposed technique: **a** The *black temporal curvilinear dotted line* represents the first skin incision performed to microsurgically isolate and prepare the donor vessel (i.e., parietal branch of the STA). The craniotomy needed to perform the STA-MCA bypass with EDMS is also indicated by the *circular black temporal dotted line*. The skin incision is extended 4 cm over the midline, after the completion of the procedure for unilateral MCA revascularization (see *green dotted line*). All the skin incisions are performed behind the hairline. The area for the two subsequent frontal craniotomies is also illustrated. **b** After splitting the temporal muscle and performing a craniotomy over the Sylvian point, the dura is opened in a star-fashion way (see *dotted lines*). The donor vessel remains intact. **c** The dural flaps are inverted on the cortex around the bone window (see *dotted lines*) in order to obtain the EDS. Thereafter, the STA-MCA bypass procedure is completed. **d** After having performed EMS (by covering the exposed cortex with the temporal muscle and by suturing the muscle to the dural edges), the bone flap is repositioned, the skin incision is then extended 4 cm over the midline behind the hairline. The scalp flap is then reflected anteriorly, and the vascularized frontal pericranial tissue (in *yellow*) is dissected and reflected on the scalp flap. Two symmetric bilateral frontal parasagittal craniotomies are performed; the underlying frontal dura is opened in a star fashion, and the dural flaps are inverted on the cortex around the frontal bone window, to obtain bifrontal EDS (see *dotted lines*). **e** To obtain bilateral frontal EPS, the pericranial flap is placed over the cortex surface and sutured to the dural edges bifrontally. The bone flaps are then repositioned. **f** This coronal section shows: the three performed craniotomies (one temporal, two frontal parasagittal); the STA-MCA bypass; the dura inverted on the cortex around each bone window (to obtain EDS); the temporal muscle laying on the exposed temporal cortex (to obtain EMS) and sutured to the dural edges; the pericranial flap placed over the frontal cortex surface (to obtain EPS) and anchored to the dural edges.

and the skin incision closed in two layers taking care not to compromise the STA-MCA bypass.

Results

Case series

Patient and operation characteristics, occurrence of complications and 30-day follow-up data are summarized in Table 1.

Between December 2009 and March 2012, we operated on eight pediatric moyamoya patients (six girls; two boys) with the combined one-staged technique (Table 1). Mean age was 10.0 years (range 4.2–17.5). Six children were affected by moyamoya disease (MMD). Of the two patients with moyamoya syndrome (MMS), one patient had neurofibromatosis type I (case number 5) and the other trisomy 6 and a 17q25 translocation (case number 6). All patients were operated on by the same senior surgeon (L.R.) at the University Medical Center, Utrecht, The Netherlands.

The mean duration of the procedure (from incision to closure of the skin) was 6 h and 54 min (range 5:10–9:35).

According to our protocol, we performed blood transfusions depending on the hemoglobin and hematocrit levels. In two children, we administered transfusions perioperatively (cases 5 and 7). In three other children, one-time blood transfusion was administered within 5 days postoperatively (cases 1–3). The blood loss perioperatively did not exceed 500 cc in any patient.

Two patients experienced complications. One patient (case number 2) had a 30-min episode of focal seizures with twitching of the left corner of the mouth 7 h postoperatively that was treated with Oxcarbazepine. CT scan showed diffuse edema in the treated right hemisphere, which resolved on follow-up neuroimaging. The other patient (case number 8) had short lasting symptoms (each lasting less than 10 min) on the fifth operative day consisting of sensibility disorders in his right arm and dysarthria. These symptoms were accompanied by aggressive behavior, which he did not experience before. These symptoms partly resembled his pre-surgical events (before anti-epileptic drugs were prescribed) and were attributed to either TIAs or focal seizures. Valproic acid was restarted and symptoms resolved within 2 days. MRI revealed no new findings.

At follow-up, no wound problems occurred and a good cosmetic outcome was achieved in all eight children.

Illustrative case

An 8-year-old previously healthy (case 3, Table 1) boy presented with multiple transient ischemic attacks (TIAs) over the span of 1 year. The TIAs consisted of symmetric leg weakness, dysphasia, right-sided hemiparesis, and episodes of migraine, mostly following agitation and hyperventilation. The neurological examination showed no deficits. DSA revealed bilateral narrowing of the internal carotid artery termination including the M1 and A1 segments with presence of bilateral compensatory collaterals confirming MMD (Fig. 3, left panel). The H_2O-PET scan showed evidence of decreased baseline CBF and impaired CVR to acetazolamide in both MCA territories and in the frontal areas bilaterally (Fig. 3, right panel).

Based on the symptomatic course of the disease, the characteristics of the TIAs and the impaired CVR in the left MCA territory and bifrontal areas, we treated the patient with the described technique (Figs. 1 and 2).

The postoperative course was uneventful. An early MRI examination at day 9 postoperatively did not reveal new ischemic lesions and MRA showed good patency of the bypass. Three TIAs were reported at 2-month follow-up (which resembled preoperative events in terms of duration and aspect). Thereafter, no further ischemic episodes were reported and the boy remained asymptomatic for the next 10 months. At 6-month follow-up, MRI/MRA showed no new ischemic lesions, stable of MMD and patency of the bypass. Repeat H_2O-PET after 1 year showed improvement of CBF and CVR in the left MCA territory and in the frontal areas bilaterally (Fig. 3, right panel).

Table 1 Patient and operation characteristics, occurrence of complications and 30-day follow-up

Case no.	Sex	MMD/MMS	Age (years) at surgery	Preoperative events	Side surgery	Perioperative complications <30 days*	Operating time (h;min)	Days of admittance
1	F	MMD	4.2	Infarction L hemisphere; TIAs R hemisphere (<10 min)	Left (+BF)	None	05:30	14
2	F	MMD	4.9	TIAs R hemisphere (<1 h)	Right (+BF)	7 h postoperatively: partial seizures R hemisphere (CT: edema,no infarction; oxcarbazepine prescribed)	07:53	10
3	M	MMD	8.4	TIAs L hemisphere and symmetric leg weakness (min–1 h); migraine	Left (+BF)	None	05:10	11
4	F	MMD	7.9	TIAs L hemisphere (min-1 h)	Left (+BF)	None	06:23	8
5	F	MMS	9.7	TIAs R hemisphere (min–1 h); migraine	Right (+BF)	None	05:33	9
6	F	MMS	9.7	Collapses (10–30 min); bilateral TIAs and infarction (recent L)	Left (+BF)	None	07:40	7
7	F	MMD	9.6	Severe cognitive impairment; dyskinesia R hand	Left (+BF)	None	09:35	9
8	M	MMD	17.5	Stuttering infarction and parzial seizures L hemisphere (temporarily valproic acid prescribed)	Left (+BF)	6 days postoperatively: 2 TIAs/partial seitures L hemisphere (<10 min; MRI no new findings; valproic acid restarted)	07:28	6

BF bifrontal, *F* female, *h* hours, *L* left, *M* male, *min* minutes, *MMD* moyamoya disease, *no*. number, *R* right

Discussion

We report a case series consisting of eight pediatric patients treated with a single-staged direct STA-MCA with EDMS for unilateral revascularization of MCA territory, in combination with bifrontal EDPS for bilateral frontal revascularization [3]. The indication to perform the one-step combined revascularization procedure was to have hemodynamic compromise and clinical symptoms involving concurrently both a MCA territory and the bifrontal areas. No significant complications occurred.

Adequate CBF supply in the bifrontal areas is of importance, especially in pediatric patients with MMV [3, 8, 9, 11, 15–18]. Cerebral ischemia in this region can in fact lead to lower extremity motor weakness and to intellectual and neuropsychological dysfunction [11, 17–21]. Although little is known about the natural history of MMV, steep decline of neurocognitive performance has been described in 44 % of

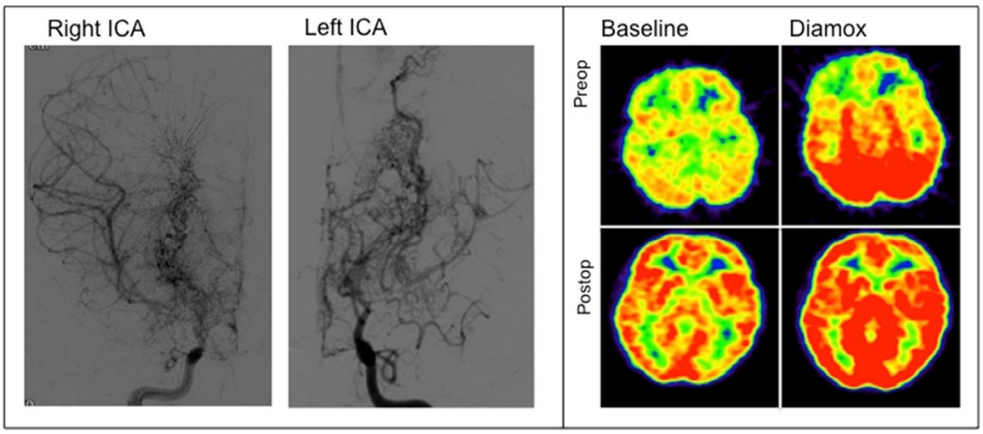


Fig. 3 *Left panel*: DSA images (anteroposterior view) showing narrowing of the ICAs termination including the M1 and A1 segment. Extensive hypertrophic lenticulostriatal collaterals are evident bilaterally. *Right panel*: Preoperative H_2O-PET before and after Diamox showing bilaterally the evidence of decreased CBF and hemodynamic failure in MCA territory and frontal areas bilaterally. One-year postoperative H_2O-PET documenting a clear hemodynamic improvement in bifrontal areas and left MCA territory

the pediatric population [20]. Furthermore, there is growing evidence that decreased CBF, especially in the frontal lobes, is correlated with diminished intelligence [16]. Most surgical approaches for MMV focus on increasing the blood supply to the MCA territory [2, 3, 13, 22]. In theory, by surgically restoring blood flow in the bifrontal areas, there may be a beneficial effect on neurocognitive performance [12]. A recent study analyzed neurocognitive profiles pre- and postoperatively on 65 pediatric patients with MMV operated by means of a combination of indirect bypass procedures (unilateral encephalo-duro-arterio-synangiosis—EDAS—in 12 patients; bilateral EDAS in 11 patients, bilateral EDAS and bifrontal encephalo-galeal-periosteal-synangiosis in 42 patients). This study demonstrated a retained intelligence quotient (IQ) and a significant improvement in performance IQ after surgery [20]. The benefits of bifrontal revascularization on long-term cognitive outcome in children with MMV, however, remain to be established in larger clinical trials.

Advantages and disadvantages of the proposed technique

The main advantage of this technique consisted of the possibility to revascularize in one-session three different vascular regions: the MCA territory unilaterally and the frontal areas bilaterally. The technique combined direct and indirect revascularization procedures: direct STA-MCA bypass increased flow immediately and EDMS promoted progressive neoangiogenesis over time in the MCA territory. Bifrontal EDPS aimed at inducing progressive neoangiogenesis over the frontal lobes bilaterally.

This technique also offered the advantage of revascularizing both frontal areas using two separate parasagittal frontal craniotomies, located 2 cm away from the midline. This reduced the risk of injuries to the SSS and the parasagittal veins. Similarly, this technique avoided exposure and opening of the interhemispheric fissure (IF) and of the arachnoid membrane of the medial frontal lobes. Furthermore, the technique allowed inverting and reflecting dural flaps under the craniotomy edges expanding thereby the cortical coverage area for neoangiogenesis.

The scalp was not incised separately for this combined procedure. The skin incision was located behind the hairline and required no shaving. The use of a single skin incision (either curvilinear or in a zigzag fashion) and three craniotomies gave an excellent cosmetic result. Large craniotomy flaps or multiple burr holes might lead to less favorable cosmetic outcomes.

Finally, the proposed technique did not compromise eventual future contralateral MCA territory revascularization, as well as revascularization procedures in the posterior circulation territory.

EDPS may represent a very useful alternative to the existing indirect procedures for frontal lobe revascularization or to direct STA-ACA bypass for revascularization of frontal areas [11]. STA-ACA bypass is known to be technically challenging, especially in pediatric patients. In fact, the site of micro-anastomosis into the ACA territory needs a very distal preparation of the STA frontal branch, and the cortical recipient of the ACA is generally also very small and often located in a sulcus. Furthermore, performing a direct STA-ACA bypass may be difficult in combination with a direct STA-MCA bypass.

The EDPS is technically easier and therefore should be feasible in most children with MMV. The use of frontal pericranial flaps to induce neoangiogenesis in patients with MMV has shown to be an effective technique [6, 11, 15, 23–26]. The choice of using the periosteum (frontal pericranium) for the bifrontal revascularization relies on the abundant blood supply, providing nourishment to the bone, and potentially promoting neoangiogenesis [11]. The frontal pericranium receives blood supply mainly from the supraorbital and supratrochlear arteries (as well as from frontal branches of the STA), emphasizing the importance of preparing the pericranial flap pediculated anteriorly over the orbits to preserve its vascular supply [25, 27].

Although the one-stage combined revascularization technique represented a long surgical procedure, the immediate postoperative and short-term (within 30 days) follow-up of the reported patient series indicated the feasibility and safety of this technique. Data on long-term clinical, neuropsychological, radiological, and hemodynamic follow-up of the whole case series is currently being collected.

The presented method for surgical revascularization in pediatric patients with MMV represents a modification of existing techniques [11, 15, 23]. Bifrontal EDPS by itself could also be used as a supplementary procedure in patients who already underwent previous revascularization procedures, in case of bifrontal hypoperfusion or progression of the MMV with symptoms referable to frontal lobe hypoperfusion. Bifrontal EDPS itself is doable by means of an incomplete bicoronal incision, from one superior temporal line to the contralateral one.

Conclusions

The novelty of this STA-MCA bypass with encephalo-duro-myo-synangiosis combined with bifrontal encephalo-duro-periosteal-synangiosis is the one-stage approach, combining direct and indirect revascularization techniques in three different vascular regions: the MCA territory unilaterally and the frontal areas bilaterally. Direct STA-MCA bypass and EDMS in the MCA territory achieves both an immediate flow augmentation and promotes neoangiogenesis over time; bifrontal EDPS aims at inducing neoangiogenesis on the frontal lobes bilaterally. Surgical risk is minimized by avoiding exposure of the superior sagittal sinus and opening of the interhemispheric

fissure. In our series, no significant complications due to surgery expansion were encountered.

Furthermore, bifrontal EDPS itself could be a valuable procedure in patients who already underwent other cerebral revascularization procedures and who present symptoms referable to frontal lobe hypoperfusion.

Acknowledgments The authors thank Mr. Peter Roth (Department of Neurosurgery, University Hospital Zurich, Zurich, Switzerland) for the drawings in Fig. 2.

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